PARALYSIS OF ALL FOUR LIMBS AND OF ONE SIDE OF THE FACE, WITH DISSOCIATION OF SENSATION, DEVELOPING IN A FEW HOURS AND RESULTING FROM MENINGO-MYELOENCEPHALITIS[1].

By Charles K. Mills, M.D.,

CLINICAL PROFESSOR OF NERVOUS DISEASES IN THE UNIVERSITY OF PENNSYLVANIA; NEUROLOGIST TO THE PHILADELPHIA HOSPITAL.

AND

By William G. Spiller, M.D.,

ASSISTANT CLINICAL PROFESSOR OF NERVOUS DISEASES AND ASSISTANT PROFESSOR OF NEUROPATHOLOGY IN THE UNIVERSITY OF PENNSYLVANIA; NEUROLOGIST TO THE PHILADELPHIA HOSPITAL.

From the William Pepper Clinical Laboratory (Phœbe A. Hearst Foundation).

We are familiar with acute syphilitic meningo-myelitis chiefly as disease of the thoracic cord. Out of twelve cases tabulated by Williamson[2], in ten the lesion was wholly or almost wholly thoracic. In one it was thoracic and lumbar, in the other it was thoracic and cervical. The cases which have fallen under our observation have been thoracico-lumbar chiefly. It may be said that the localization to one portion of the cord is seldom absolute, in other words, although the lesion may be situated chiefly in one region of the cord, as the thoracic, some infiltration of round cells, and other evidences of meningo-myelitis are usually found in other regions. Cervical or cervico-bulbar forms of acute syphilitic meningo-myelitis, which might be designated clinically as high myelitis, are almost as rare as high tabes compared with the tabes which attacks the cord at lower levels, but in this cervical form the lesions are not confined to the cervical region, and are merely most intense in this portion of the spinal cord. Syphilitic meningo-myelitis is often associated with meningo-encephalitis. The following case is therefore in the first place of interest because of the unusual location of the myelitis. It also has other points of interest, as the manner in which the facial nerve and its nucleus on one side were attacked, its simulation of hemorrhage into the cervical portion of the spinal cord, and the interesting sensory and reflex phenomena.

---

[1] Read at the annual meeting of the American Neurological Association, June 5, 6 and 7, 1902.

The patient, a colored man, forty-five years old, a laborer, was admitted to the Men's Nervous Wards of the Philadelphia Hospital, January 28, 1902. He had had gonorrhea twice, and a sore on the penis two years before. He had been a hard drinker. His family history was unimportant.

On September 26, 1901, the patient stated that he fell from a wagon, striking on his shoulder. He immediately rose, climbed to his wagon and continued working. Two weeks later he was obliged to quit work on account of pain in his left arm. He went to the Philadelphia Hospital for treatment, where he remained for about three weeks, at the end of which time he was again able to work. He worked until December 25, 1901, when pain in his left arm and between his shoulders compelled him to discontinue. The pain lasted for about one month.

At about 3 P.M., January 22, 1902, he suddenly lost the use of his left arm, this paralysis being followed in a short time by loss of control of the left leg; later by paralysis of the right side of the face, of the right arm and then of the right leg. Five or six hours elapsed between the time when he first noticed paralysis of the left arm and the time when the right leg became paralyzed.

He complained of swelling and of dull pain in the parts after they were paralyzed, but more especially in the left arm. He felt no pain immediately preceding the paralysis, and he was not unconscious at the time. He was not aphasic. After the paralysis he had a severe headache which ceased only the day before his admission to the hospital.

On admission (January 28, 1902), his general appearance was that of a well developed and well nourished colored man. He had had no movement of his bowels for five days before admission, and had voided no urine for at least twelve hours. Examination showed right facial paralysis, including the frontalis and orbicularis palpebrarum muscles. When the eyebrows were elevated no wrinkles were produced on the right side of the face. He was unable to fully close the right eye, and lachrymation was present. Speech was muffled, the mouth being drawn to the left side when attempts were made to talk, and whistling was impossible. Paralysis of the sixth nerve was not apparent. Hearing on the left side had been affected since the patient was eight or nine years old, and he could not hear the tick of a watch close to his ear; on the right he could hear the tick at about ten or twelve inches. Tinnitus aurium was not present. He had no difficulty in swallowing, but complained of difficulty in keeping food in his mouth when eating. The uvula was not deflected to either side. His sense of taste was preserved, a solution of quinine and spirits of camphor and sweetened water being recognized on the tip and right half of the tongue. The tongue deviated slightly to the left. The pupils were contracted, the left irregular in outline; irides reacted sluggishly to light and on accommodation. Hemianopsia was not apparent. Knee-jerks and Babinski reflex were absent; the grip on both the right and left side was zero.

His chest appeared normal, the clavicles were large and prominent, and the supraclavicular fossæ depressed. The lungs were resonant anteriorly. Examination of the heart revealed a systolic murmur; his pulse was rapid, full and easily compressed. His abdomen was normal in appearance and was not distended.

The patient was again examined on January twenty-ninth. His breathing was labored, and his voice weaker than on the day previous. He complained of pain in his shoulder. On the thirtieth of January his breathing was less labored. He did not talk much, and answered by nodding his head. It was reported that he passed blood with his stools. His appetite was good and he had slept well the night previous.

Between January twenty-ninth and February first he was visited several times by both of us. The results of our examinations are summarized below, his general condition not changing much during this period.

The patient was fully conscious, and had no aphasia. His voice was a mere whisper, but he understood all that was said to him. He said positively that he had not been unconscious even during the development of the paralysis, and he was equally positive that he had not had convulsions. Wrinkles had disappeared on the right side of his forehead. Bilateral ptosis, equal on both sides, was present, and could not be overcome by voluntary efforts to open his eyes. When the eyelids were closed distinct paresis of the right orbicularis palpebrarum was present, the right palpebral fissure being about 5 cm. in width. In showing his teeth his mouth was drawn to the left side, the right side of his face remaining immobile. The tongue protruded very slightly toward the left, even when the right corner of the mouth was drawn up by the hand of the examiner; the masseter contracted firmly; the uvula was in the median line, and no distinct paralysis of the soft palate was present. The pupils were equal; both irides responded to light and promptly in convergence. The eyeballs exhibited nothing abnormal. Movements of the head from side to side and from before backward could be freely made.

The patient could shrug his shoulders well, but with the exception of this movement paralysis of both upper extremities was complete. The limbs were flaccid, no contracture being present. Both lower limbs were paralyzed in all parts. The ophthalmic (supraorbital) reflex was present on the left, absent on the right. The conjunctival reflex was normal on the left, and greatly diminished on the right. Jaw-jerk was slightly exaggerated. The coraco-brachial or inner shoulder reflex was present. Biceps-jerk was almost lost on each side; the triceps-jerk was preserved on each side. The wrist-reflex was lost on each side. Knee-jerk was present on each side, but diminished, a little prompter on the right. The ankle-jerk was present on each side, and slightly exaggerated on the right. Ankle-clonus and the Babinski reflex were absent on each side.

The sensory conditions of this patient were of unusual interest. Tactile sense was preserved on both sides of the face, in the upper extremities and on both sides of the thorax and abdomen. Temperature sense (tested for heat and cold), and pain sense, were preserved on both sides of the face and neck, but were lost on the right side of the chest and abdomen, and diminished on the left side of the same. Temperature and pain senses were also lost in the distal portions of the right upper extremity. At a point about three inches above the elbow, hot, cold, and painful impressions began to be recognized. The pain and temperature senses were diminished in the left upper extremity. Sensation for touch was fully preserved in the left lower extremity, and somewhat diminished in the distal portion of the right lower extremity. Sensation for heat, cold and pain was almost completely lost in the left lower extremity, and completely lost in the right.

Respiration was entirely costal in type. The heart-beat was very rapid; a loud systolic murmur was present, loudest at the second intercostal space on the left side; the second sound of the heart was accentuated. The murmur was fully as distinct in the second intercostal space on the right side as in the corresponding space on the left. He had had no disturbance of bowels or bladder previous to his paralytic attack. When examined on January twenty-ninth and thirtieth he had incontinence of urine and feces; catheterization was necessary. He had had no dizziness nor vomiting at any time preceding the paralysis. He stated that his sight had been poor for three months. No ophthalmoscopic examination was made.

On February first he was somewhat duller, and early in the day did not seem to know where he was. On the next day his condition was much the same, but his pulse and respiration were more rapid, and he complained of pain between the shoulders. On the third of February the patient frequently protruded his tongue to moisten his lips, and complained of his mouth being dry. He was duller and weaker than on the previous day, and did not pay so much attention to questions. His respirations were shallow. He had not slept well the night before. He refused milk, but asked occasionally for water; his appetite was fairly good. On February 4, 1902, at 2.30 A.M., he died.

A summary of the most important coarse pathological appearances found on necropsy is as follows: Edema of the spinal cord in the upper cervical region, adhesion of the dura: chronic splenitis and perisplenitis, old fibrous splenic adhesions; calcified tuberculous nodules of the right lung. A detailed account of the examination of the brain is subjoined.

The calvarium was thick, measuring at the occipital end 1.25 cm. and at the frontal end 1 cm. The dura was tightly adherent to the calvarium and a small new growth was present directly in front of the pituitary fossa, not extending above the surface of the dura. This growth had two distinct parts. A small nodule was present in the middle cerebral fossa (left), and was soft to the touch.

The brain was somewhat edematous in the parietal and occipital lobes. There was no indication of hemorrhage in the brain or pons, and no hemorrhage in either lateral ventricle was apparent. The fluid in the lateral ventricles was not increased in amount.

No distinct abnormalities of the choroid plexus were observed. Horizontal section of the upper portion of the basal nuclei revealed no pathological condition. No gross lesions were found in the brain. The masses noted in the base of the skull did not extend into the bone itself, but lay within the dura.

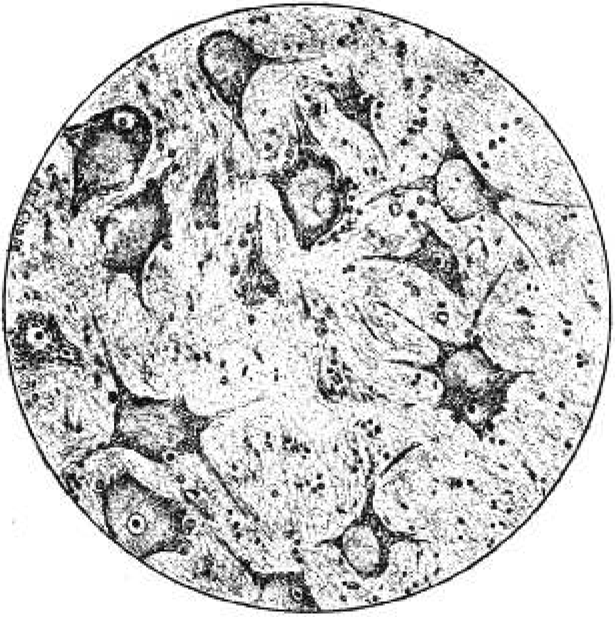
An increased amount of cerebrospinal fluid was found between the meninges and cord.

The brain and spinal cord were removed and examined microscopically.

Sections of the brain and spinal cord were stained by the Weigert hematoxylin and Marchi methods, by thionin, hemalum and acid fuchsin. Intense round-cell infiltration was found in the pia over the optic commissure, both in the walls of the vessels of the pia, and about some of the blood vessels within the commissure. The optic nerves and commissure as shown by Weigert's hematoxylin stain, were not degenerated.

Intense round-cell infiltration was found in the pia covering the medulla oblongata and the walls of some of the pial vessels were thickened. Here and there within the medulla oblongata a small vessel was found with round-cell infiltration about it.

Some of the cell-bodies of the left facial nucleus were degenerated, *i.e.,* the nucleus was displaced to the periphery of the cell-body and the chromophilic elements were broken up, but most of the cell-bodies of the left facial nucleus were normal or nearly normal. The right facial nucleus was exceedingly degenerated, and in most of the cell-bodies the chromophilic elements at the center of the cell-body had become granular or had disappeared, and only a few of these elements remained at the periphery of the cell-bodies; in others, no chromophilic elements at all were found. The nuclei were displaced to the periphery of the cell-body. The contrast afforded by the two facial nuclei was very striking. The intramedullary portion of the right facial root was deeply stained by the Weigert hematoxylin and did not appear to be degenerated by this method. The degeneration of the facial nerve was evidently of very recent origin, and it was impossible to say whether the disease of this nerve began in its nucleus or in its peripheral portion. The alteration of the cell-bodies was such as could be produced by disease of the peripheral portion of the nerve. The round-cell infiltration in the pia over the medulla oblongata was not greater on



Nerve cell-bodies of the right facial nucleus seen in one field of the microscope. The cell-bodies are drawn in their relative positions as shown by the microscope, and almost all are intensely degenerated.

the right side than on the left, so that the degeneration of the right facial nerve could hardly be explained by the meningitis. The nerve may have been injured in its peripheral portion by a small growth like those described within the dura of the base of the skull; and the paralysis may have existed longer than the patient was aware of.

The fourth, fifth and sixth cervical segments were so softened by inflammatory changes that the normal relations of white to gray matter were entirely altered, and a large portion of a transverse section at this level was not colored at all by the Weigert hematoxylin stain, because of degeneration of the medullary substance. The round-cell infiltration within the spinal cord and pia at this level was intense. Here and there were swollen axones or spaces from which the axones had disappeared. Some of the small blood vessels within the spinal cord and pia at this level had much thickened walls, and numerous very small hemorrhages were found within the spinal cord. Some of the vessels of the pia were almost entirely closed by the intense proliferation of the intima.

The round-cell infiltration within the pia was very intense at the eighth cervical segment, but considerably less so than at the area of softening. The veins of the pia were thickened, and in some of them round-cell infiltration was intense, especially within the inner coat. The small arteries of the pia at this level were not intensely thickened. No degeneration at the eighth cervical segment could be found by Weigert's hematoxylin method. The direct pyramidal tracts were found degenerated when the Marchi method was employed, but elsewhere at the eighth cervical segment the degeneration as shown by the Marchi method was very slight. Slight degeneration was found by this method in the left column of Burdach near the posterior horn, and may have been caused by degeneration of the posterior roots of this segment. The destruction of nerve fibers at the area of softening had not existed sufficiently long to cause much secondary degeneration detectable even by the Marchi method. Most of the cell-bodies of the lower cervical region, as shown by the thionin stain, were normal or nearly normal. Collections of small bacilli were found within the spinal cord at the eighth cervical segment. It seems hardly probable that these bacilli were the cause of the alteration of the central nervous system, and they may have invaded the spinal cord shortly before or shortly after death. The intense thickening of the blood vessels at the area of softening indicated that the process had been of long standing, and that the softening was probably the final result of a long-existing impaired circulation. The changes were such as are seen in syphilis, and the case probably was one of syphilis.

Round-cell infiltration was found in the pia of the mid-thoracic region, but was not quite so intense as in the eighth cervical segment. No degeneration by Weigert's hematoxylin stain was found at the mid-thoracic region.

The round-cell infiltration of the pia in the lumbar region was about as intense as in the mid-thoracic region. Some of the cell-bodies of the anterior horns of the lumbar region were tumefied and much pigmented.

In this case the rapidity with which the paralysis of the four extremities and of the face, with sensory, pulmonary and other symptoms of a serious character, developed made the diagnosis of spinal hemorrhage into the cervical region or into the oblongata-spinal transition seem probable, and this diagnosis was at first made. The pathological examination showed acute myelitis of high grade, the foundations of which were laid in previous disease of the vessels and membranes. Numerous small hemorrhages were present in the more or less inflamed and disintegrated cervical cord, but these were secondary, or at the most concomitant with the attack of myelitis. In severe acute myelitis hemorrhages almost invariably occur, these in some instances being punctiform, in others of considerable extent, even sufficient to cause some bulging of the cord; not infrequently both punctiform hemorrhages and those of larger size are present in the same case. The fundamental disease in these cases is, however, the myelitis, although this usually occurs on the basis of previous disease of the spinal vessels.

When the question of diagnosis is between myelitis and hemorrhage, this is understood to mean between primary hemorrhage into the spinal cord (hematomyelia) and myelitis, or between primary hemorrhage into the spinal membranes (hematorrhachis) and myelitis. The manner of onset may give the clue to the diagnosis between hemorrhage into the cord and acute myelitis. In hemorrhage the attack is usually sudden and without premonitory symptoms, although this rule is not without rare exceptions, as when a paralysis preceded by some paresthesia comes on slowly. The development of paralysis in the case here recorded was relatively rapid; nevertheless five hours were occupied in its completion. Prodromal symptoms were not entirely absent, even presuming that the pain in the arm which followed the fall of four months previous had no connection with the attack of myelitis, it will be remembered that for one month previous to the development of paralysis in the four extremities the man had had pain between his shoulders and in his left arm. Other prodromal symptoms were absent, but this was sufficient to cast some doubt upon the diagnosis of primary hemorrhage into the cord. The usual causes of hemorrhage were absent, unless the fall of four months previous may have been thought to bear some causal relation to the attack, but the traumatism was too remote to have induced a gross spinal hemorrhage. It was more likely to play a rôle in the determination of an inflammatory process in the spinal cord of a syphilitic subject. The patient had suffered from syphilis, the common cause of acute myelitis.

The diagnosis from spinal meningeal hemorrhage, extradural or subdural, was less difficult than that from primary hematomyelia. In spinal meningeal hemorrhage the symptoms of irritation come on suddenly and are usually of a very severe type, although even this rule is not without exceptions, probably in cases in which the hemorrhage into the spinal spaces occurs gradually. A meningeal hemorrhage sufficient to have produced the profound paralytic, sensory, reflex and other symptoms which were present in our case, would in all probability have been ushered in by severe pain in the back and in the distribution of the nerves whose roots were irritated by hemorrhage, and in addition by rigidity of the back, possibly local spasm in nerve distributions, contractures and even more or less generalized convulsions. With the exception of a steady pain in the back and one arm these irritative phenomena were not exhibited by the patient prior to the paralysis. In spinal meningeal hemorrhage paralysis usually occurs after the phenomena of irritation have been well marked, while in both acute myelitis and hematomyelia, whether primary or secondary, the paralysis is one of the earliest symptoms. The usual causes of spinal meningeal hemorrhage were absent in our case, while as already stated, syphilis, the common cause of myelitis, acute or chronic, was present. The course taken by the disease was not that of spinal meningeal hemorrhage, which usually results in death because of the extent of the inundation, or in partial recovery through absorption after the lapse of considerable time. The rapid development of the paralysis of all the limbs resulting from myelitis makes the case one of unusual interest.

Another disease from which a case like the one here recorded might need to be differentiated is spinal or cerebrospinal tuberculous meningomyelitis. A pure spinal tuberculous meningitis is rare, but the cerebrospinal affection is not uncommon, and when the lesions are basal and spinal, the latter predominating, the case may show some features similar to those presented by a syphilitic cervical or cervicothoracic myelitis of the acute or at least of the subacute type. A case has been reported by Hensen[3] in which the diagnosis of cerebrospinal syphilis was made, but in which the autopsy showed the case to be one primarily of tuberculous meningitis, inflammation and degeneration of the spinal cord taking place secondarily. This case is in itself one of unusual interest, and is worthy of being recorded in detail in this connection.

A woman forty years old became sick eight days before she was received into the hospital with chill and severe headache. These symptoms were soon followed by severe pain in the back and the region of the kidneys, especially on the right side. A few days later disturbances in micturition occurred, requiring catheterization. On the eighth day of her disease she was taken into the hospital with a diagnosis of kidney and bladder disease. On admission she was mentally disturbed, confused and excited. She had pains in her head and back, in the region of the kidneys and in the legs. The atlanto-occipital articulation was tender to pressure, but no rigidity of the neck was present. There was, however, extraordinary rigidity of the whole vertebral column. No vertebra, however, was especially tender. Slight abducens paresis on the right side; bladder distended, detrusor paralysis; some hyperesthesia in the legs; patellar reflex exaggerated; other reflexes normal. The patient with support and even alone was able to take a few steps, although with rigid back and legs. No choked disc, not even later. The statement that out of 12 children she had borne most of them had died in early life aroused the suspicion of syphilis, although no other symptoms of syphilis were obtained. Some improvement occurred in the patient's condition, the mental excitement diminished, and voluntary urination became possible. Temperature was diminished. On the other hand, however, rigidity of the neck and of the vertebral column became constant. The hyperesthesia also diminished. On the eighteenth day of the disease indisputable symptoms of spinal cord disease in the thoracic region developed. She complained of paresthesia in the right leg and then in the left. She was unable to raise the lower limbs. The muscles became flaccid, and marked reflex contractions developed in testing for the patellar reflex and in irritation of the skin. A few days later girdle pain in the abdomen developed on each side as high as the eighth rib; the abdominal reflex was lost. On the twentieth day of the disease the legs became completely paralyzed, and sensation in the lower parts of the body was almost lost. The patellar reflex was lost, the muscle excitability in the peroneal muscles on both right and left side was increased. In the left anterior tibial muscle there was beginning reaction of degeneration. Incontinence of feces and tendency to decubitus. Now cerebral symptoms again became conspicuous. The sensorium became clouded. The patient became somewhat delirious at night, and there was some evidence of facial paralysis which sometimes seemed stronger on the right side, sometimes stronger on the left. The abducens paralysis which at first had come on had disappeared. The muscles of the eye were not implicated. On the twenty-first and twenty-second days the mental disturbance developed into profound stupor, and slow forced movements appeared in the upper limbs, and only occasionally seemed to be somewhat coördinated. The diagnosis was made of cerebrospinal syphilis.

At the necropsy the diagnosis of syphilis was not confirmed, but a basal tuberculous meningitis with more extensive tuberculous meningitis of the spinal membranes, especially in the thoracic region, and secondary softening of the cord were found. Also miliary tuberculosis in the lungs, liver, kidneys and ulcers in the vermiform appendix.

The microscopical examination showed that the pia-arachnoid was in a state of inflammation throughout the entire length of the spinal cord, but more especially in the thoracic region. The inflammatory condition was more pronounced on the posterior aspect than on the anterior. The walls of the vessels, arteries as well as veins, were thickly infiltrated by round cells, almost difficult to recognize, and their layers could not be distinguished. Some vessels were thrombosed. The small-cell infiltration extended from the vessels into the surrounding pia. Distinct tubercles were not found, possibly on account of the short duration of the disease, but the absence of tubercles is not uncommon in tuberculosis of the cerebral membranes. However a few typical giant cells were present. In the middle thoracic region only scattered portions of the gray matter could be recognized, in which a few ganglion cells could be discovered after considerable searching. By the Pal stain a few normal nerve fibers were found, most of them, however, had disappeared. No tubercles were found in any part of the spinal cord substance.

From the pathologico-anatomical point of view the degeneration of the spinal cord was secondary to the meningeal disease, especially of the vessels of the meninges—everywhere where there was meningeal disease of high grade there was degeneration in the border zone of the spinal cord also, whereas in the thoracic region there was complete destruction and dissolution of the spinal cord substance. The primary lesion, however, was the vascular lesion of the meninges.

A comparative study of Hensen's case and of the case here recorded shows that in the case of tuberculous meningo-myelitis the paralytic, sensory and reflex symptoms of grave type developed gradually and after a period in which the symptoms of meningeal irritation were prominent. It was eighteen days before indisputable symptoms of involvement of the cord were exhibited, and twenty days before paralysis was complete. The extensive meningeal inflammation was shown by the rigidity of the back, and also probably by the hyperesthesia of the extremities. In our case paralysis of all four limbs was complete in five hours, and the grave sensory, reflex and visceral phenomena came on with almost equal rapidity. With the exception of pain between the shoulders and in the left arm, symptoms of meningeal inflammation were absent. Basal symptoms were present in both cases, but most of these developed in the tuberculous case late in the disease, while the facial paralysis in our case was a prominent feature in the paralytic syndrome which came to the surface in the short period of five hours if the patient's statement may be relied upon.

It is well to add that the diagnosis of tuberculous meningomyelitis in Hensen's case was made from the condition of the lungs and intestines, and not from the presence of tubercle bacilli within the spinal cord.[4]

---

[2] "Syphilitic Diseases of the Spinal Cord," Manchester, 1899.

[3] Hensen, H., Deutsche Zeitschr. f. Nervenheilk., v. 21, No. 3-4, p. 240.

[4] A case of tuberculous meningomyelitis with paralysis has been published by Joseph Collins in the December, 1902, number of this JOURNAL.